Adult Wilms' Tumor With a Unique Presentation of High-Grade Fever, Photophobia, and Headache

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Wilms' tumor is the second most common tumor in children, accounting for 6% to 7% of all childhood tumors. However, in adults, it is a rare occurrence. The true incidence of adult Wilms' tumor is difficult to ascertain because of its rarity in the adult population. A review of literature demonstrates that fewer than 300 cases have been reported worldwide. Treatment guidelines in adults have not been established, although reported prognosis is worse for adults compared with children because the disease is more advanced in adults at the time of diagnosis. Reported here is a case of adult Wilms' tumor presenting as high-grade fever and abnormal laboratory values.

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KEY WORDS

Wilms' tumor • Renal mass • Open radical nephrectomy • Multimodal therapy

The patient, a 26-year-old woman, presented to the emergency department complaining of 105.8°F fever, generalized body aches, photophobia, headache, and neck stiffness. She also had epigastric discomfort along with one episode of emesis. She denied any lower urinary tract symptoms or history of renal calculi. Her past medical and surgical histories were unremarkable. She had no family history of urologic disorders or malignancies. She denied dysuria but did notice a decrease in her urine output over the previous 2 days. Physical examination was unremarkable and she had no flank tenderness. Laboratory examination was also unremarkable with the exception of her creatinine

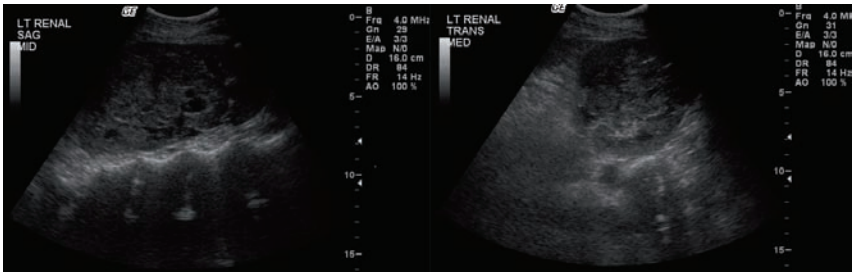

Figure 1. Ultrasound images of questionable renal mass.

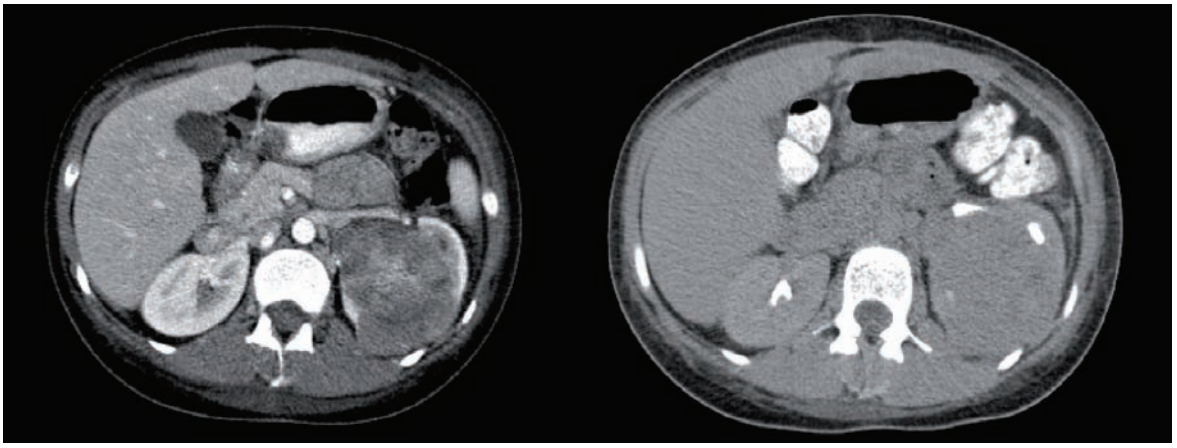

Figure 2. Computed tomography images of solid mass in the left kidney.

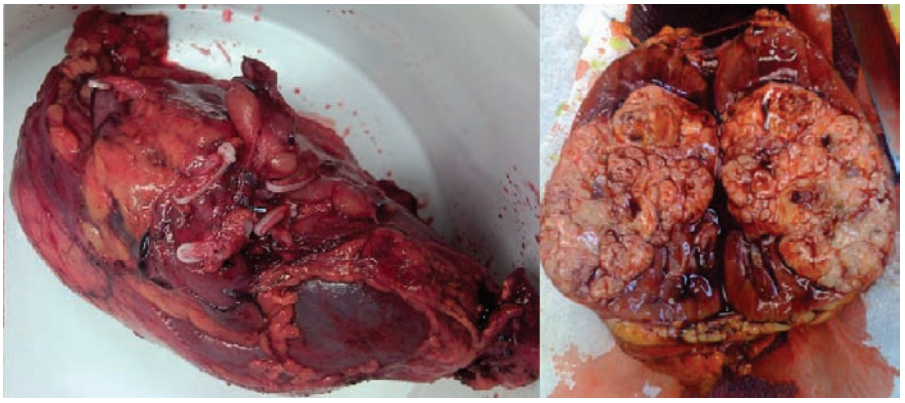

Figure 3. Nephrectomy specimen.

level, which was 2.09 mg/dL. Urinalysis demonstrated moderate blood and an erythrocyte count of 3 to 5 $\times$ 10^9/L. She was admitted to the general medical floor with the diagnosis of meningitis. Renal ultrasound demonstrated a questionable renal mass (Figure 1), and computed tomography (CT) demonstrated at 12-cm solid mass in the left kidney (Figure 2). Chest imaging and bone scanning results were both negative. She immediately underwent an open radical nephrectomy (Figure 3).

On gross inspection, the tumor measured 9 $\times$ 7 cm and was a gray-tan firm nodule subjacent to the renal capsule. Sectioning of tumor showed several small areas of focal hemorrhage, possible necrosis, and a multinodular appearance. Renal parenchyma away from the tumor area showed pale gray focal discoloration. Microscopic analysis revealed that the tumor was a nephroblastoma without anaplasia. The blastema area stained positive for vimentin and pan-cytokeratin (PAN-CK). The epithelial components were strongly positive for PAN-CK and focally positive for vimentin. Wilms' tumor gene protein staining was positive throughout. Pathologic staging was pT2 with primarily undifferentiated blastema and epithelial tubular structures (Figure 4). All lymph nodes were negative for tumor.

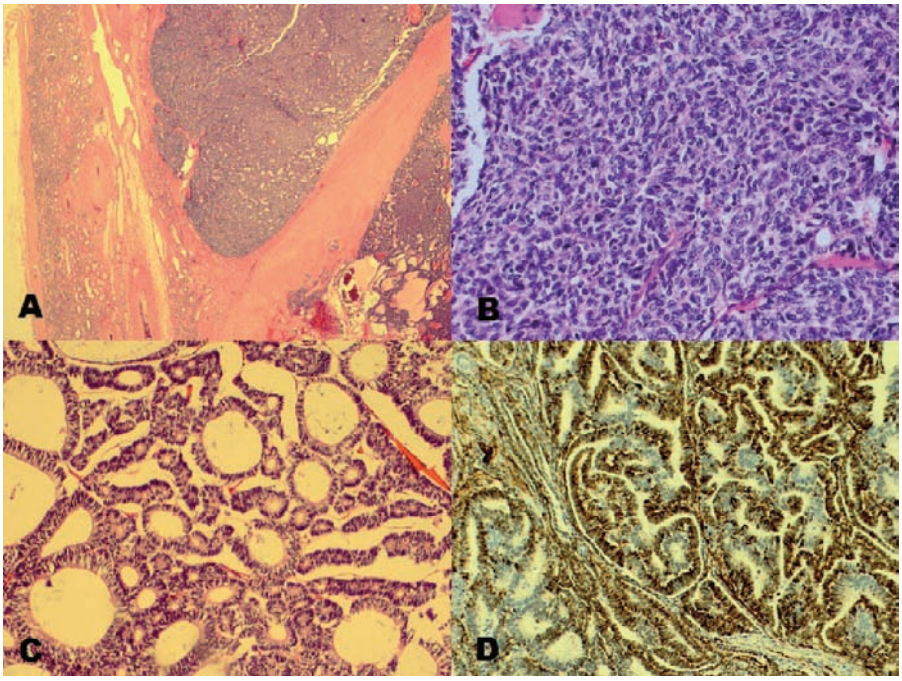

Figure 4. Pathology images of nephroblastoma. **(A)** Presence of epithelial, stromal, and blastemal elements. **(B)** Evidence of immature spindle cells. **(C)** Glomerulotubular structures. **(D)** Immature tubular structures.

Postoperatively, the patient was treated for 22 weeks with vincristine and actinomycin D, which she tolerated with minimal side effects. Her 1-year follow-up imaging demonstrated no disease.

Discussion

Wilms' tumor is the second most common tumor in children, accounting for 6% to 7% of all childhood tumors; however, in adults, it is a rare occurrence. The true incidence of adult Wilms' tumor is difficult to ascertain because of its rarity in the adult population. A review of literature demonstrated that fewer than 300 cases have been reported worldwide. Treatment guidelines in adults have not been established although the reported prognosis is worse for adults compared with children because the disease is more advanced in adults at the time of diagnosis.

In 1980, Kilton and colleagues developed the following criteria for the diagnosis of adult Wilms' tumor: presence of a primary renal neoplasm, presence of a primitive blastematous spindle or round-cell component, formation of abortive or embryonal tubular or glomeruloid structures, absence of tumor diagnostic of renal cell carcinoma (RCC), and age $>$ 15 years.[1]

The diagnostic criteria for Wilms' tumor include the presence of abortive or embryonic glomerulotubular structures within an immature spindle-cell stroma. These structures are not found in RCC. The absence of fetal renal tissue points against the diagnosis of Wilms' tumor.[2] The most common location of nephrogenic blastema is the subcapsular portion of the renal cortex,[3] which explains this neoplasm's common appearance as an exophytic mass.

Microscopically, both adult and childhood Wilms' tumors have epithelial, stromal, and blastemal elements.[3] Typically, adult Wilms' tumor usually presents in young patients as a large, rapidly growing abdominal mass, but the presentation may vary. The oldest patient reported in the literature with a confirmed diagnosis of adult Wilms' tumor was age 84 years.[4] More common in women, approximately 20% of patients are diagnosed between the ages of 15 and 20 years, and 80% are diagnosed between the third and seventh decades.[5]

Abdominal CT imaging typically shows a large, well-defined, exophytic, inhomogeneous mass that is cortical in origin. There may be large areas of low density present along with areas of focal necrosis and hemorrhage on noncontrast images. After intravenous contrast administration, there may be variable enhancement, and the remaining normal parenchyma will appear as a pseudocapsule around the tumors (75%).[6] Ultrasonographic imaging typically demonstrates a large, complex mass with large cystic components compared with RCC, which is usually a heterogeneous solid mass.[7]

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Although there are no defined treatment protocols for adults, many authors suggest using

childhood protocols for the treatment of adult Wilms' tumors; the pathologic staging used is also the same for pediatric patients.[8]

In 1982, the National Wilms' Tumor Study (NWTS) group reported 31 adult Wilms' tumor patients who were treated with multimodal therapy between 1968 and 1979. The 3-year survival rate was 24% compared with 74% in childhood patients. The authors concluded that adult Wilms' tumor has a worse prognosis compared with Wilms tumor occurring in children and should be treated aggressively with three-drug chemotherapy and radiotherapy to the tumor bed (4500 cGY), regardless of the stage of disease.[9]

The 1990 NWTS report described 27 adults treated between 1979 and 1987 and found the 3-year survival rate with multimodal therapy 67% when anaplastic tumors were included, and 79% when they were excluded.[10]

In 2003, Reinhard and colleagues reviewed 30 adult patients who were treated with surgery and chemotherapy, as well as 14 out of 30 patients receiving irradiation. They reported a 4-year median overall survival rate of 83%.[11]

Conclusions

Unfortunately, due to the rarity of adult Wilms' tumor, sample sizes are limited and firm conclusions cannot be made regarding specific treatment protocols. After diagnosis, a multimodal therapeutic approach must be explored with the cooperation of the urologist and the oncologist.[12-15]

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MAIN POINTS

- Wilms' tumor is the second most common tumor in children; however, in adults it is a rare occurrence. Fewer than 300 adult cases have been reported worldwide. Treatment guidelines in adults have not been established although the prognosis is worse for adults compared with children because the disease is more advanced in adults at the time of diagnosis.
- Criteria for diagnosis of adult Wilms' tumor include presence of a primary renal neoplasm, presence of a primitive blastematous spindle or round-cell component, formation of abortive or embryonal tubular or glomeruloid structures, absence of tumor diagnostic of renal cell carcinoma, and age $>$ 15 years.
- Although there are no defined treatment protocols for adults, many authors suggest using childhood protocols for the treatment of adult Wilms' tumors; the pathologic staging used is also the same for pediatric patients.
- Due to the rarity of adult Wilms' tumor, sample sizes are limited and firm conclusions cannot be made regarding specific treatment protocols. After diagnosis, a multimodal therapeutic approach must be explored with the cooperation of the urologist and the oncologist.